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Case Report

A Rare Case of Synovial Sarcoma Presenting in Paraspinal Region

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ABSTRACT

Synovial sarcoma is an aggressive malignant neoplasm arising from mesenchymal tissue representing 7-10% of all soft tissue sarcomas. Typically, it presents in children and young adults aged 13-35 years of age. These are usually located in the extremities, most commonly around the knee. Paraspinal region is a rare site of synovial sarcoma and thus requires a high index of suspicion for timely diagnosis. Here, we discuss such a rare case of paraspinal synovial sarcoma.

Key words: Soft tissue sarcoma, Paraspinal, Synovial sarcoma, Dorsal vertebrae, Metastatic synovial sarcoma.

INTRODUCTION

Synovial sarcoma (SS) is an aggressive malignant neoplasm arising from mesenchymal tissue, that differentiates to resemble synovial cells.[1] It represents 7-10% of all soft tissue sarcomas.[2] Typically, it presents in children and young adults aged 13-35 years of age. Eighty five percent of SS arise in the extremities with a preponderance for lower extremities, mainly in region around the knee. Only 3% arise in the head and neck region.[3] Presentation in paraspinal region is very rare and often remains undiagnosed for long durations of time. Here, we present a rare case of SS arising in the lower dorsal spine in a 46 year old male.

CASE REPORT

A 46 year old gentleman presented at a local hospital with complaint of pain upper back. He was treated with painkillers, muscle relaxants and physiotherapy for six months but did not have any improvement in his symptoms. He subsequently noticed a swelling in upper back, for which Contrast Enhanced Computed Tomography (CECT) scan was done. CECT revealed a soft tissue density mass on right side of chest and upper part of abdomen starting from lower part of scapular region and measuring 7.5×8×2.5 cm. The mass was well-defined and lying in subcutaneous tissue with maintained fat planes with surrounding structures.

This mass was excised and sent for pathological assessment. Histopathological examination (HPE) revealed a malignant mesenchymal tumour with possibility of monophasic SS. On Immunohistochemistry (IHC) tumour was positive for vimentin and negative for cytokeratin (Figure 1) and thus, patient was considered as a case of monophasic SS for treatment.

In view of malignant finding on histopathology, patient was advised 18-Fluorodeoxy Glucose Positron Emission Tomography-Computed Tomography (18-FDG PET-CT) scan, which revealed FDG avid heterogeneously enhancing soft tissue thickening in right costal pleura infiltrating the posterior chest wall. There were multiple FDG-avid mediastinal lymph nodes. Liver showed FDG-avid hypodense lesions noted in both lobes of liver largest 6.9×5.9 cm in segment VIII of liver suggestive of hepatic metastasis. Spleen also showed FDG-avid hypodense lesions, largest measuring 2.9 x 2.9 cm. Another FDG-avid lesion was noted in the paraspinal region at the level of D12 vertebra with evidence of faint FDG avid soft tissue stranding. FDG avid lytic destructive changes were also noted in D8 vertebrae suggestive of metastasis (Figure 2).

As patient had extensively metastatic disease he was started on anthracycline and ifosfamide based chemotherapy. Patient had relief of pain and significant decrease in size of swelling clinically, after the first two cycles of chemotherapy and is currently under treatment.

DISCUSSION

SS typically presents in young adults as a painless mass in the extremities, usually in the knee.[3] Eighty percent of the synovial sarcomas are located in the extremities with only five percent tumours located in the spinal axis. If located near joints they may cause pain with movement and may be mistaken for bursitis or myositis. It presents in two forms monophasic and biphasic. Monophasic contains only spindle cells while biphasic contains both spindle cells and epithelial cells. The translocation (X; 18) is specifically associated with SS. This leads to fusion of SYT-SSX1 genes. According to a recent retrospec-

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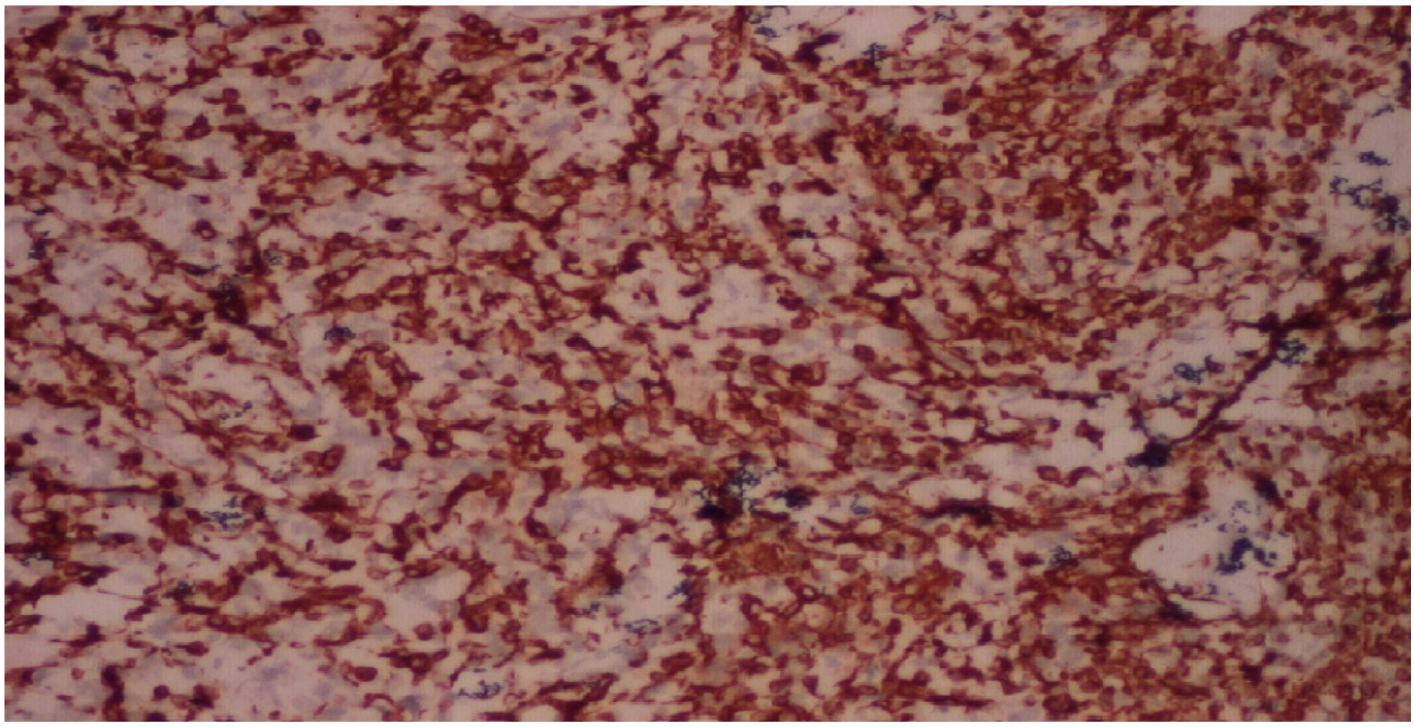

Figure 1: Spindle shaped cells demonstrating positivity for vimentin suggestive of synovial sarcoma.

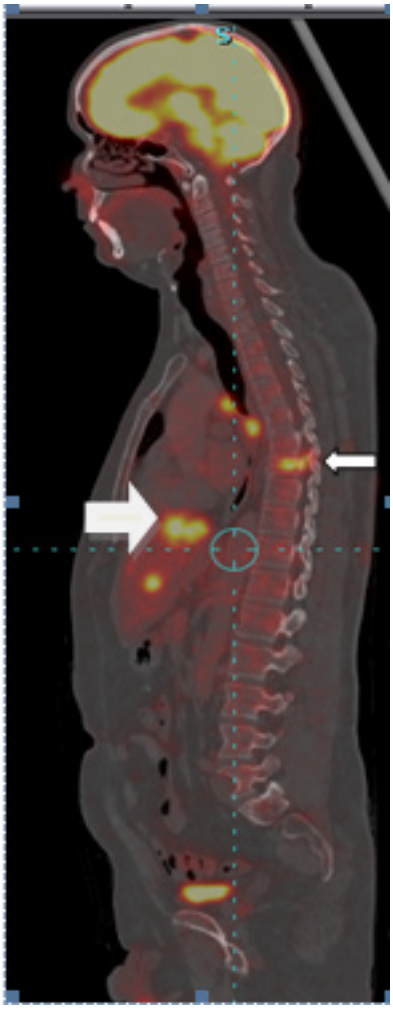

Figure 2: FDG PET-CT Scan of the patient showing FDG avid paraspinal mass(thin arrow) and multiple liver metastases (thick arrow).

tive study this fusion is associated with biphasic SS, increased proliferative rate and a poor prognosis.[4] Our patient had a monophasic type of SS, which confers a better prognosis compared to biphasic type.

Synovial tumors located in the paraspinal region may give rise to neurological deficits. Progressively increasing symptoms and size of the lesion may be an indication of a malignant process. Our patient presented with pain initially which was mis-diagnosed as myalgia and was treated accordingly for six months. Only when patient developed swelling at the site of pain, he was considered for further evaluation. By the time diagnosis was made, patient was metastatic. In a review of 2,568 patients of sarcoma by Smith *et al*[5] the mean size of lesions was 10.7 cm at diagnosis before 2000 and 9.6 cm after 2000. The mean duration of symptoms was 26 weeks for soft tissue sarcomas. According to a study by Grimer[6] a 1 cm increase in size of lesion reduces the chances of cure by 3-5%. To prevent a delay in diagnosis, an updated guidance for such conditions was introduced with the concept of two-week-wait period.[7] According to this any swelling greater than 5 cm in size, painful, deep to deep fascia, progressive and recurrent after excision should be considered malignant unless proven otherwise. Thus, a high index of suspicion should be kept for patients not responding to initial management and should be evaluated early to rule out more grave conditions.

Radiologically, in 50% of patients with synovial sarcoma, plain radiographic findings are normal. Approximately, 30% of patients have calcifications on plain radiography or CT which are typically diffused punctate and often more concentrated at the periphery than at the center of the mass.[8]

Magnetic Resonance Imaging (MRI) provides superior tissue characterization. It can aid in preoperative planning, assist in grading the tumor and assessing clinical prognosis. Thus, MRI is the investigation of choice.

On T1-weighted images most tumors display heterogeneous intermediate signal intensity, secondary to extensive areas of hemorrhage and necrosis. However, small lesions mostly display homogenous intensity and may be mistaken for benign lesions. On T2-weighted images, the tumors are usually hyperintense, with a signal intensity similar to, or lower than, that of fatty tissue; fluid-fluid levels are demonstrated in the cystic components in 10%-25% of tumors.[8] Approximately one third of lesions, typically high grade, demonstrate a triple signal-intensity pattern on T2-weighted images which consists of mixtures of hyperintense fluid, intermediate fat like signal intensity and hypointensity similar to fibrous tissue.[9]

Differential diagnosis of this patient consisted of metastatic disease to spine, plasmacytoma, lymphoma and leukemic infiltration, however, histological findings and IHC clinched the diagnosis of SS.

Treatment of localized SS consists of surgery followed by radiation or chemotherapy. Chemotherapy forms the mainstay of treatment of metastatic SS. It has relatively high response rate to chemotherapy, this may be largely due to the fact that synovial sarcoma patients are younger than patients with other subtypes. Thus, these patients have better performance status and are able to tolerate chemotherapy better and are more likely to complete treatment without complications. Ifosfamide based chemotherapy has shown significant benefit in this subtype of sarcoma.[10]

Management remains largely the same for paraspinal SS except for the limited possibility of surgical resection due to the complex anatomy of the region. In this patient excision biopsy was done to establish the diagnosis, this was followed by chemotherapy as the patient had extensively metastatic disease on FDG-PET CT scan.

CONCLUSION

Paraspinal synovial sarcomas are rare. A high index of suspicion forms the key to early diagnosis and curative therapy.

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CONFLICT OF INTEREST

None

ABBREVIATIONS

SS: Synovial Sarcoma; CECT: Contrast EnhancedComputed Tomography; IHC: Immunohistochemistry; 18-FDG-PET-CT: 18 Flourodeoxy Glucose Positron Emission Tomography Computed Tmography; MRI: Magnetic Resonance Imaging; CT: Computed Tomography.

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